Subacute Diencephalic Necrosis and Dural Arteriovenous Malformation

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We report a case of congestive venous necrosis involving the diencephalon symmetrically, presumably precipitated by a dural arteriovenous malformation (AVM). The patient presented with a 1-month history of intermittent confusion and gait ataxia. The initial radiological work-up revealed a dural AVM. The patient's subsequent deterioration was accompanied by computed tomographic findings of diencephalic congestion. Pathological examination showed subacute necrosis of the diencephalon. We discuss the pathogenesis of this case of subacute diencephalic necorsis and its possible relationship to the entity of subacute diencephalic angioencephalopathy. (Neurosurgery 17:653–656, 1985)

Key words: Arteriovenous malformation, Diencephalon, Sinus thrombosis, Subacute diencephalic angioencephalopathy

Pure dural arteriovenous malformations (AVMs) are rare and may be congenital or acquired (1, 3, 5). The symptoms are usually related to the abnormal circulation or a pressure effect, e.g., a subjective buzzing noise, audible bruits, cranial nerve weakness, or the cavernous sinus syndrome. Subarachnoid hemorrhage and seizures, the most common symptoms in pial or mixed (pial and dural) AVMs, are rare in pure dural AVMs (3, 5, 6). We report a case of subacute diencephalic necrosis presumably precipitated by a dural AVM and discuss its relationship to subacute diencephalic angioencephalopathy.

CASE REPORT

A 63-year-old man was admitted with a 1-month history of intermittent confusion, disorientation, and gait ataxia. Four days before admission, the patient was noted to have hallucinations and episodes of urinary and fecal incontinence. His medical history was significant for hypertension, angina, mild congestive heart failure, and colonic polyps for which he had undergone a partial colectomy.

On admission, he was alert but mildly confused. He was oriented to self and time, but misnamed the place. He could follow three-step commands and recall three of three items at 5 minutes. His speech was normal. The cranial nerves were intact. The findings of his motor and sensory examinations were unremarkable. He showed mild truncal and gait ataxia. The rest of the cerebellar examination was unremarkable. His deep tendon reflexes were normal. His plantar responses were flexor bilaterally. A complete blood count, electrolyte determination, liver function and renal function were all normal. Toxic/drug screening was negative. The stool was positive for occult blood. His cerebrospinal fluid examination was normal. An electroencephalogram showed a mildly slowed background. Computed tomography (CT) on the 7th day of hospitalization revealed a vascular malformation in the left occipital area (Fig. 1). Plain x-ray films of the skull showed prominent vascular markings of the posterior branch of the middle meningeal artery, suggesting dural involvement. A barium enema on the 8th day of hospitalization revealed a large 2- \times 3-cm polyp in the proximal transverse colon and two small 5- \times 5-mm polyps in the sigmoid colon. Colonoscopy and biopsy were performed on the 11th day of hospitalization. The pathological diagnosis of the large polyp was villous adenoma. Angiography obtained on the 18th day of hospitalization showed a dural AVM fed by the posterior branch of the middle meningeal artery and draining into the deep venous system through a large anomalous vein in the occipital area. The straight sinus was not visualized (Fig. 2). The dural AVM was thought to be an incidental finding. However, the patient's condition slowly continued to worsen. During the following week, he became apathetic and grew progressively more lethargic. Subsequently, he became comatose with occasional jittering. CT on the 23rd day of hospitalization showed diffuse low attenuation of the diencephalon with mild mass effect (Fig. 3).

It was hypothesized that the dural AVM was causing venous congestion of the diencephalon. Therefore, under general anesthesia, a small craniotomy was performed and the dural feeding artery was ligated. The draining veins of the AVM were not exposed or ligated. However, the patient remained

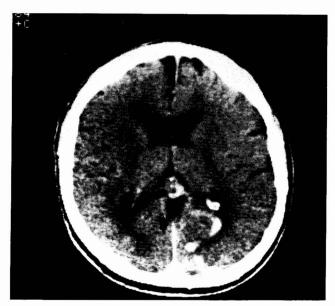


FIG. 1. CT on the 7th day of hospitalization shows a vascular malformation in the left occipital area. The straight sinus is not seen (arrow).

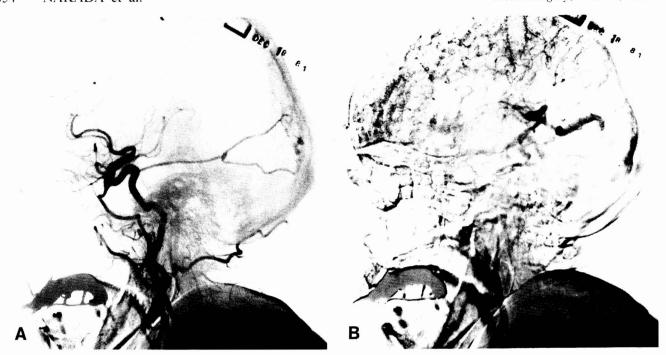


Fig. 2. Cerebral angiography on the 18th day of hospitalization. A, arterial phase shows a dural AVM fed by the posterior branch of the middle meningeal artery. B, intermediate phase shows a large anomalous vein draining into the deep venous system. The straight sinus is not visualized.

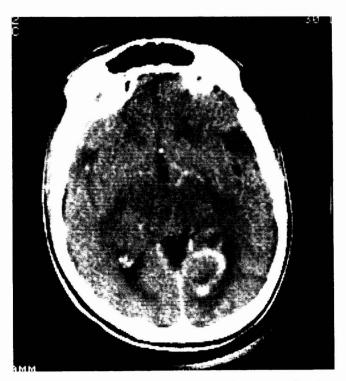


Fig. 3. CT on the 23rd day of hospitalization shows diffuse low attenuation of the diencephalon with mild mass effect.

comatose and died on the 44th day of hospitalization. Autopsy was limited to the brain.

Fresh gross examination revealed postsurgical changes of the dural AVM. The major venous sinuses, including the straight sinus, were patent. After formalin fixation, the gross pathological examination showed a large venous channel in the subarachnoid space of the left occipital pole. However, no abnormal vessels or hemorrhage was seen in the occipital lobes except for several splinter hemorrhages at one level. Microscopic examination showed a moderate amount of recent hemorrhage immediately adjacent to the wall of the straight sinus. Its lumen, however, was patent (Fig. 4). In the thalamus, subacute necrosis (aproximately 3 to 4 weeks old) was indicated by the presence of focal collections of lipid- and hemosiderin-filled macrophages in the center, microcysts, and gemistocytic astrocytes in the relatively intact periphery (Fig. 5). Scattered throughout the thalami, cortex, and white matter of the occipital lobes bilaterally were numerous large caliber vessels measuring up to 2 mm in diameter. These vessels had thin collagenous walls devoid of elastica and muscularis. Many of these vessels contained organized thrombi (Fig. 6). The arteries showed mild atherosclerotic changes, but no occlusion was found.

DISCUSSION

Failure to visualize the patient's straight sinus by angiography on the 18th day of hospitalization is a good indicator of disturbed flow in the straight sinus. The hemorrhagic foci adjacent to the straight sinus and the numerous thrombotic, thin-walled anomalous vessels in the diencephalon noted on pathological examination are further support of the existence of disturbed flow in the straight sinus. Therefore, our patient's bilateral diencephalic congestion and necrosis very likely resulted from the high volume blood inflow from the dural AVM, which was possibly impounded by impaired venous drainage into the straight sinus.

In 1974, DeGirolami and colleagues reported a case of progressive dementia due to bilateral diencephalic necrosis (2). At autopsy, the primary pathological changes were seen in the vessels and consisted of necrosis, fibrinoid deposits, collagenous proliferation, and eventual obliteration of the lumina. This pathological process was seen in both the arterial

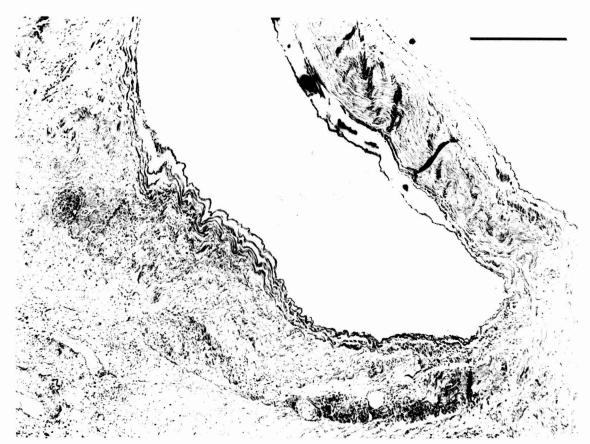


Fig. 4. Photomicrograph of a section of the straight sinus shows an intact endothelial lining and a patent lumen. Bar = 0.5 min. (Hematoxylineosin stain. \times 52.)

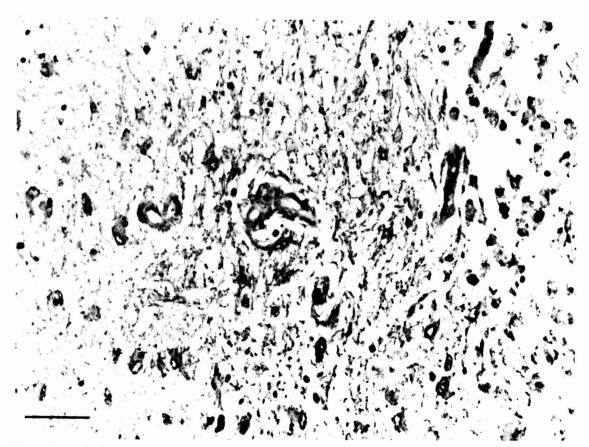


Fig. 5. Photomicrograph shows an infarct in the left thalamus with lipid-filled macrophages in the central cystic portion at the *upper right* and fibrillary astrocytes on the cyst periphery. $Bar = 50 \mu m$. (Hematoxylin-eosin stain, \times 325.)

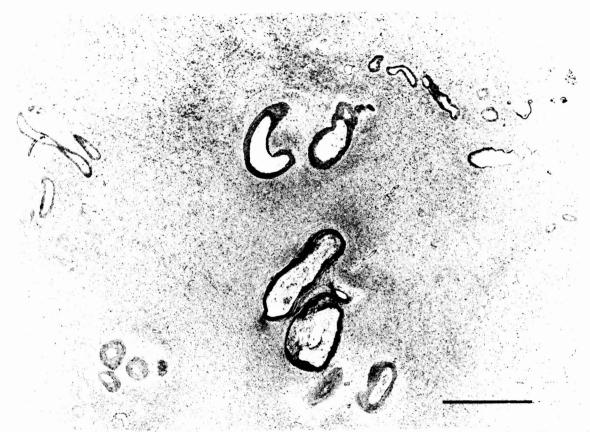


Fig. 6. Photomicrograph shows thrombosed large diameter veins in the right thalamus. Bar = 1 mm. (Hematoxylin-van Gieson stain. \times 23.)

and the venous vessels. Although a mononuclear inflammatory infiltrate was seen, the cellular exudate was far less abundant than that encountered in the various types of vasculitis. Bilateral carotid arteriograms were reported to show no abnormalities, but no information regarding the venous phase and the straight sinus was given. DeGirolami et al. coined the term "subacute diencephalic angioencephalopathy" to describe this unique case. Six years later, Kinney and his colleagues reported a similar case of subacute diencephalic angioencephalopathy (4). Again, the primary pathological finding at autopsy was confined to the diencephalon and was characterized by patchy parenchymal necrosis and proliferative vascular change. In this case, there was clear involvement of the venous system with proliferative change in the internal cerebral veins, their large feeding veins, and the veins lining the 3rd ventricle. Although no complete occlusion of deep cerebral veins was found, the authors concluded that the pathological findings of their case may be best explained by occlusion of the deep venous system. Angiographic findings were not described in their report.

The similarities of the clinical and pathological findings of our case and the two cases of subacute diencephalic angioencephalopathy are striking except for the presence of dural AVM. The angiographical findings of our case demonstrated the presence of disturbed flow in the straight sinus. Given that our patient had subacute diencephalic angioencephalopathy clinically and pathologically, it follows that subacute diencephalic angioencephalopathy may indeed represent a disease process of the deep cerebral venous system resulting in disturbed venous flow and congestion of the diencephalon, as suggested by Kinney and his colleagues.

Our patient's diencephalic necrosis was very likely attributable to the high blood inflow due to the dural AVM

exacerbated by the impaired flow of the straight sinus. Unfortunately, ligation of the dural feeding artery in this case did not improve the patient's clinical condition, although earlier surgical intervention might have had a more favorable clinical effect. Clinical awareness of the possibility of a treatable condition due to an otherwise benign form of dural AVM is warranted.

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